

Abstracts

Out of Town Meeting of the Ulster Paediatric Society, Saturday 17th May 2008

Shandon Hotel, Dunfanaghy, Ireland.



PROGRAMME:

- 11.00am Welcome – President: Dr Denis Carson
- 11.10am Invited Guest Speaker
- 11.45am Spoken Presentations
- 12.30pm Poster Discussion
- 12.40pm Annual General Meeting
- 13.00pm Close

PRESENTED ABSTRACTS

1. First febrile convulsions – unnecessary hospital admissions?

Armstrong K, Dalzell E.

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Background: In Northern Ireland all children with first febrile convulsion are admitted to hospital. Reasons for admission vary from historical, to parental anxiety and concern regarding underlying clinical diagnosis. This practice varies from current practice throughout the UK and Ireland¹. This audit looks at admissions over a one-year period with a view to changing practice.

Aims: To look at all hospital admissions with febrile convulsion over a one year period in order to review current practice, study patient outcomes and to make decisions regarding future management protocols.

Methods: Retrospective case note analysis of all children coded as presenting with febrile convulsion to the emergency department, RBHSC in the time period 1st September 2003 to 31st August 2004 were obtained. Clinical information was gathered from current care pathway.

Results: 155 children were admitted with first febrile convulsion. There was a slight male predominance with the median age of admission 20 months. There was a positive family history in 30%. The median temperature on arrival was 38.7 degrees Celsius. Blood sugar was recorded on arrival in 92% of cases with median BM 6.6. Median seizure duration was 5 minutes with 72% described as tonic clonic in nature.

57% of children had bloods taken during the admission

with MSSU being performed on 86%. Anticonvulsants were required in 12% of cases prior to or on arrival to A&E. 98% of children received antipyretics, 41% were discharged home on antibiotics. Median admission duration was 1 day with URTI being the commonest diagnosis (65%) There were no cases of meningitis or septicaemia. There were no PICU admissions and no deaths.

Conclusions: Admission policies will be reviewed and an alternative care pathway derived following this study. Results indicate that it is not necessary to admit all children presenting with first febrile convulsion. Children who have a confirmed clinical diagnosis and in whom there is no parental anxiety and adequate education could be discharged home after a period of observation. There were no cases of meningitis or septicaemia presenting as first febrile convulsion with the most common underlying diagnosis being URTI.

1. Evidence based Guideline for post-seizure management in children presenting acutely to secondary care, Baumer HJ. *Arch Dis Child* 2004;89:278 – 280.

2. An unusual cause of vaginal bleeding in a pubertal girl

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Clear cell adenocarcinoma of the cervix is extremely rare in childhood with no reported cases in the National Registry of Childhood Cancer in the last 25 years. We describe the case of an 11yr old girl with clear cell adenocarcinoma, to highlight genital tract malignancies as an important differential in the investigation of vaginal bleeding in children.

An 11yr old pubertal girl presented with persistent vaginal bleeding, having had regular menses since menarche two years previously. Hormone therapy for 3 months did not improve her symptoms and she underwent an EUA. This revealed a mass in the cervix, subsequently identified histologically as a clear cell adenocarcinoma of the cervix. MR imaging showed local spread. She received chemotherapy and radiotherapy, including brachytherapy with some tumour response and remains under regular review.

Vaginal bleeding is the commonest presenting symptom of genital tract malignancies in the paediatric population¹ and warrants prompt investigation. Despite this, children often present with advanced disease¹. An EUA should be

considered early in the investigation of vaginal bleeding, as it can lead to earlier tumour detection and subsequent improved prognosis.

1. McNall RY *et al.* Adenocarcinoma of the Cervix and Vagina in Pediatric Patients. *Pediatr Blood Cancer* 2004;43:289-294.

3. Audit of clinical management and treatment of patients with hypopituitarism presenting in infancy

N Abid, D Carson

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Objective: To determine the clinical features present in infancy, age of presentation, diagnostic strategies and treatment of congenital and idiopathic hypopituitarism.

Method: The data were collected retrospectively, from 1978 to 2008, on all the patients with features of hypopituitarism present in infancy diagnosed at the Endocrine unit of Royal Belfast Hospital for Sick Children. Data included age at presentation, gender, clinical features, neuroradiology findings (MRI, CT and USS), endocrine deficiency, hormone replacement therapy and outcome.

Results: Total of 31 patients were studied, with clinical features of hypopituitarism in infancy. 6 patients had SOD with intact septum pellucidum, 9 with septo optic dysplasia with absent septum pellucidum, 8 with agenesis of corpus callosum, 2 with holoprosencephaly, 1 with schizencephaly and 6 with isolated pituitary hypoplasia. The mean age of referral to endocrinology unit was 1.43 with a range of 0 to 7.23 years, with a female to male ratio of 1.2:1. The most common feature present in infancy was prolonged jaundice and ocular abnormality followed by hypoglycaemia. 2 patients each had hypernatraemia and hyponatraemia. Out of 14 male patients 6 had micropallus and 2 had undescended testes. Seizures were present in 5 patients. 3 patients had cleft lip and palate, 2 had microcephaly and 1 each had macrocephaly, single incisor and upper limb deformity. Most of patients had multiple pituitary hormone deficiency with growth hormone, Thyroid stimulating hormone and Adreno corticotrophic hormone deficiency occurring most commonly followed by ADH and LH/FSH. Appropriate hormone replacement therapy was given at varying ages including 6 male and 1 female treated with sex hormones for delayed puberty. There was no mortality but a significant morbidity was noted including developmental delay in 19, blindness in 16, behavioural problems in 5 and sleep disturbances in 3 of these patients. We compared our data with a similar study conducted at Department of Endocrinology, Great Ormond Street Hospital for Sick Children in 1999 and found our results to be similar in terms of clinical features, pituitary dysfunction and morbidity.

Conclusion: Features of congenital hypopituitarism are nearly always present in infancy and recognition of these features can lead to early diagnosis and treatment which may improve the outcome.

4. Audit of the assessment and management of children presenting to the Emergency Department of the Royal Belfast Hospital for Sick Children over a five year time period with a diagnosis of Bell's palsy.

Julie-Ann Maney,

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Background: Bell's palsy is an acute peripheral facial nerve paralysis that usually affects only one side of the face. The aetiology is unknown; viral infection, vascular ischaemia and autoimmune disorders have all been considered as possible mechanisms. Corticosteroids and anti-viral medication have been used to treat the condition. The evidence for using these in children is sparse. Steroids are used as it is believed to decrease the inflammation and oedema of the nerve sheath. Acyclovir has been used as it thought to be due to herpes simplex infection.

Following a recent case presentation of Bell's palsy and literature review we reviewed the assessment, management, treatment and outcome of children with Bell's palsy.

Method: Retrospective audit of the assessment, management and outcome of children presenting with Bell's palsy

Retrospective audit using the symphony computer system in the Emergency Department to identify children with a presenting complaint or diagnosis of Bell's palsy, Facial palsy, or facial weakness. These case notes were reviewed individually and using an audit proforma, diagnosis, treatment and management were noted. The cases were then contacted by telephone for the outcome information. Excel was used as a database.

Results: 48 cases were identified and audited. All children had a full neurological examination and the diagnosis was clearly made. The results show wide variations in treatment; from no treatment, steroids alone, to steroids and anti-viral treatment. Most children were referred and seen by an ear, nose and throat specialist. Outcomes varied in the length of time for recovery.

Discussion: Bell's palsy is an uncommon condition. Given the natural history of spontaneous recovery in paediatric patients and given the lack of scientific evidence, the use of steroids and anti-viral medication is not evidence based at present. A large randomised controlled trial is required to establish the benefit if any of steroids and anti-viral medication. Even in our department treatment options varied widely and consensus of treatment needs to be reached in the paediatric community.

5. Is better metabolic control achieved by increasing the frequency of insulin injections?

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Introduction: The Diabetes Control and Complications Trial (DCCT) demonstrated that intensive insulin regimens (multiple-injection or pumps) delayed the onset and slowed the progression of diabetic complications in 13 to 17 year olds compared to those treated conventionally (one or two-injections daily). Those intensively treated achieved improved glycaemic control (HbA1c) but had a twofold increase in obesity and a threefold increase in severe hypoglycaemic

episodes¹. Conversely studies have shown deteriorating HbA1c levels in children changed to an intensive insulin regime². In the RBHSC Diabetic Clinic a large proportion of patients have changed to three or four (basal bolus) -injection regimes to improve control, for greater social flexibility, in preparation for the adult transition clinic or to decrease the incidence of severe hypoglycaemic episodes.

Aims: To measure changes in HbA1c, Body Mass Index (BMI) and incidence of severe hypoglycaemia after changing to 3 or more injections daily and to compare to a group who have remained on twice daily injections.

Methods: Data was collected retrospectively on children and adolescents using the TWINKLE database. Patients transferred to the adult clinic in were followed using the Diamond database. Statistical comparison was performed.

Results: 95 patients were on a 4 injection regimen with an age range of 8-21 years. 84 were on a three-injection regimen with an age range of 5-21 years. A control group of 50 patients on a twice daily regimen were identified, age range of 5-16 years. Differences in HbA1c, BMI and number of severe hypoglycaemic episodes at baseline (before regimen change) will be discussed in comparison to data 1 and 2 years later.

Conclusions: Glycaemic control over time has not improved despite large numbers of patients changing to intensive insulin regimes, especially in the adolescent years. ISPAD and NICE recommendations for young people with type 1 diabetes to have an HbA1c <7.5% are not being achieved^{3,4}. The benefits perceived by healthcare professionals of changing to intensive insulin therapy with multiple injections to improve HbA1c may be adversely affected by confounding factors such as poor compliance that our study is unable to measure. Worryingly increasing BMIs may be associated with poorer adolescent Quality of Life (QOL)⁵. Close review of the children on multiple insulin regimens will continue with intensive input from the diabetes team.

1. Diabetes Control and Complications Trial Research Group. Effect of intensive diabetes treatment on the development and progression of long-term complications in adolescents with insulin-dependent diabetes mellitus: Diabetes Control and Complications Trial. *J Pediatr* 1994;**125**(2):177-88.
2. Holl RW, Swift PG, Mortensen HB et al. Insulin injection regimens and metabolic control in an international survey of adolescents with type 1 diabetes over 3 years: results from the Hvidore Study Group. *Eur J Pediatr* 2003;**162**(1):22-9.
3. International Society for Paediatric and Adolescent Diabetes (ISPAD) Clinical Practice Consensus Guidelines 2006-2007.
4. National Institute for Clinical Excellence (NICE). Type 1 diabetes: diagnosis and management of type 1 diabetes in children and young people. 2004. Available from www.nice.org.uk/pdf/CG015childrenfullguideline.pdf.
5. Hoey H, Aanstoot HJ, Chiarelli F et al. Good metabolic control is associated with better quality of life in 2,101 adolescents with type 1 diabetes. *Diabetes Care* 2001;**24**(11):1923-8.

6. Trends in neonatal mortality over a 30 year period – what are the remaining challenges?

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¹Neonatal Intensive Care Unit, Royal Jubilee Maternity

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²Confidential Inquiry into Maternal and Child Health (CEMACH), N Ireland

Aims: A report on neonatal mortality in N Ireland during 1974 and 1975 showed it was higher than the rest of the United Kingdom. It also highlighted significant deficiencies in perinatal services. Our aim was to review neonatal mortality and the organisation of services in 2004/05 compared with the earlier study.

Methods: All babies who died before 28 days of age between 1st January 2004 and 31st December 2005 were included. Data was collected on neonatal mortality rates, causes of death, place of birth and maternity services. Cause of death was classified using mortality tabulation based on ICD10.

Results: In the 70s there were 44 maternity units and only 1 neonatal intensive care cot with 1 part time neonatologist. There are now 10 obstetric units and 19 intensive care cots, all appropriately staffed. The neonatal mortality rate during 1974/75 and 2004/5 fell from 13.3 to 4.1 per 1,000 live births. Immaturity is now the main cause of death with significantly more being less than 24 weeks (31.2% vs. 2.8%). There are notably fewer deaths primarily from RDS. (Table I). Congenital malformation remains a major cause of death but with a marked reduction in some groups such as congenital heart disease (6% vs. 26.5%) and the virtual disappearance of deaths from neural tube defects.

Conclusion: Neonatal death has fallen rapidly in 30 years due to major advances in prenatal diagnosis and neonatal therapy. The problems that remain are with very immature babies and those with sporadic lethal syndromes or other major malformations.

Table I

	1974 - 1975		2004 – 2005	
	Number	%	Number	%
Congenital malformation	219	30.9	50	26.5
Immaturity	153	21.6	77	40.7
Respiratory distress syndrome	144	20.3	12	6.3
Death from intrapartum causes	90	12.7	16	8.5
Pneumonia	28	4.0	7	3.7
Miscellaneous	74	10.5	27	14.3
TOTAL	708	100	189	100

7. The first heart valve donation from Northern Ireland to the Oxford heart valve bank

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Background: Organ and tissue donation is normally dealt with in the context of the intensive care setting with ventilated patients. In the neonatal intensive care unit organ donation is usually not possible due to the immaturity and small size of organs. There is little awareness in Northern Ireland about the

option of heart valve tissue donation. Heart valves of small sizes are needed to facilitate repair of congenital heart disease in young infants.

Aims: To promote awareness of the possibility of heart valve donation in babies. To share the experience of organising the first donation from Northern Ireland in an infant with severe chronic lung disease. To inform about the practical procedures involved in heart valve donation.

Case report: KR was born as the first twin at 27 weeks gestation (birth weight 790g) in a district general hospital. His mother had ruptured membranes for two weeks. KR required initial respiratory support and was changed to low flow oxygen on day 29. On day 43 he was transferred to the regional neonatal unit for ongoing care and management of chronic lung disease. He developed severe chronic lung disease refractory to treatment with oral/inhaled corticosteroids and sildenafil. KR deteriorated at a corrected age of eight months (weight 6 kg) and developed severe respiratory failure triggered by a chest infection. His parents decided for palliative care and against re-ventilation. His mother enquired about the possibility of KR as a potential organ donor. After consultation with the regional transplant co-ordinator and the national tissue bank the Oxford Heart Valve Bank was identified to accept heart valve tissue of such a small infant. Both parents agreed to a post-mortem to facilitate the retrieval of the whole heart and consented for tissue donation. KR died peacefully the same evening surrounded by his family. He spent the night with his parents and the post-mortem was performed the following day. It confirmed severe chronic lung disease. The heart was sent to the Oxford Heart Valve Bank, where the pulmonary and aortic valves were retrieved - both of good quality and size. These valves could be used for example for the repair of truncus arteriosus, pulmonary atresia and the Ross procedure in infants.

Conclusion: Heart valve tissue donation in small infants is possible, even if they are not ventilated. Tissue donation can offer some comfort to parents and staff, particularly if the stay in the unit was long. The regional transplant co-ordinator service is exceptionally supportive and offers good feedback to both family and staff. A flow chart on how to organise a tissue donation is now kept in the bereavement resource box of our unit and sessions for the information of staff were organised. A second heart valve donation has since taken place.

8. Did you check their calcium's? – life threatening hypercalcaemia in twin boys requiring pamidronate therapy.

Judith A Brown, Neil Corrigan

Altnagelvin Area Hospital. UK.

This case describes ten-month-old twin boys who presented with constipation and failure to thrive secondary to life threatening hypercalcaemia. They required treatment with intravenous Pamidronate.

The boys presented to the paediatric department at ten months of age with a three-week history of constipation and vomiting. Clinically they were pale and constipated with reduced muscle tone. They were failing to thrive and had been generally cross and irritable for the preceding three months.

The twins were both found to be markedly hypercalcaemic – calcium levels were 4.22mmol/l and 3.55mmol/l (normal 2.1-2.55mmol/l). Phosphate and alkaline phosphatase levels were normal.

Their calcium levels continued to rise to life threatening levels in the following twenty – four hours, peaking at 4.74 mmol/l and 3.95mmol/l.

Emergency treatment was commenced with the twins receiving forced saline diuresis and intravenous frusemide. Despite this treatment there was ongoing significant hypercalcaemia and the twins were given 0.5mg/kg of Pamidronate as a dilute saline infusion. Calcium levels reduced and four days following the pamidronate infusion levels were 2.85mmol/l and 2.47mmol/l.

Despite extensive investigation a precise diagnosis is yet to be made as to the aetiology of their hypercalcaemia however investigations thus far suggest a probable calcium receptor disorder and this will be discussed.

At review their calcium levels remain modestly elevated on a low calcium diet. There has been a marked clinical improvement in both twins following correction of their hypercalcaemia.